Paediatric Cardiology

Paediatric Syncope; Assessment, referral and management guidelines

Staff relevant to:	Children's Hospital & Neonatal staff referring to UHL paediatric cardiology service
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Contents

1. Information & who this guideline applies to:

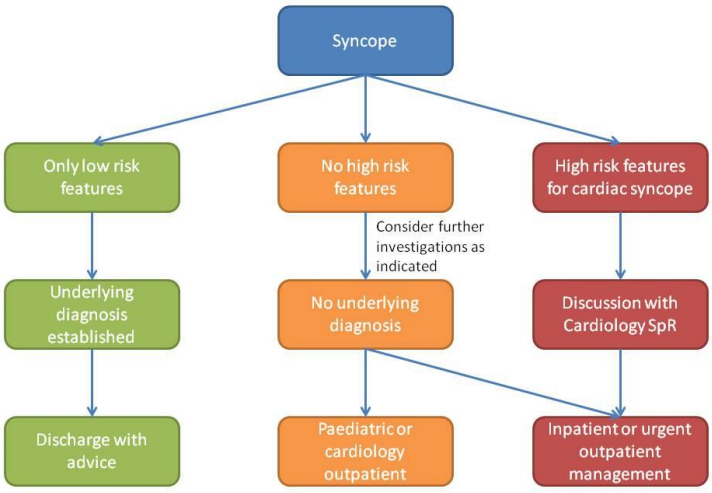
This guidance is intended to provide guidance for paediatricians and GPs on the assessment and management of the fainting child and young person. It should assist in deciding whether a child needs additional investigations and whether that can be done in primary care and general paediatrics and to emphasise which syncope has life threatening potential and requires referral to Paediatric Cardiology.

It should be read in conjunction with the Referral Guidelines for Paediatric Cardiology, our Chest pain guidelines and others yet to be created. It is not exhaustive and is likely to need revision.

It does not apply to urgent referrals for inpatients nor for those diagnosed prenatally with major congenital heart disease.
It does not replace requests for telephone advice from Cardiology consultants and Registrars (see Medirota), nor GP use of the Advice and Guidance facility.

NB: this is not meant to be entirely comprehensive and is not a substitute for a detailed professional assessment.
If there is any doubt, please discuss with a Paediatric Cardiologist.

Brief Cardiological Syncope Referral Guideline.
(For GPs also see PRISM referral questions)



MOST PEOPLE WHO FAINT DO NOT NEED TO SEE A CARDIOLOGIST

RISK ASSESSMENT

Actual Loss of consciousness Y/N
Was it witnessed; appearance (pale/flushed), video recording? .....
Duration (<30 seconds is a concern)……
Frequency; e.g. if frequent and longstanding likely lower risk; ……
Medication (prescribed or other); …….
Sensorineural deafness Y/N

Event <u>History;</u>

- of palpitations pre-syncope (defined as feeling of change in heart rate or rhythm) Y/N
- during exertion Y/N
- After exertion N/Y
- Preceded by chest pain Y/N
- On Swimming Y/N
- On sudden loud noise/shock Y/N
- With CHD (current or prior) Y/N
- With 1st degree relative sudden death / serious arrhythmia Y/N
- On prolonged standing N/Y
- On / soon after standing N/Y
- After ‘straining’ N/Y
- Related to ‘gore’ N/Y
- After shower/bath N/Y

If Yes in any of the left hand options, refer to Cardiology urgently.

If No to the entire left hand column, refer to detailed guideline, for management in primary care or paediatrics.

<u>Examination</u>

- Evidence of injury Y/N
- With a murmur? Y/N
- Bradycardic? Y/N
- Hypotensive Y/N
- Postural BP drop N/Y
- Upper-lower limb BP gradient? Y/N
- If ECG performed; Abnormal ECG? Y/N (ask for ECG review if uncertain)

2. Investigation and management of syncope

This clinical guideline focuses on investigation and management of syncope.

Transient loss of consciousness (TLOC) is defined as a state of real or apparent LOC with loss of awareness, characterised by amnesia for the period of unconsciousness, abnormal motor control, loss of responsiveness and a short duration [(1)].

TLOC groups can be categorised using pathophysiology: the qualifying criterion for syncope is cerebral hypoperfusion; for epileptic seizures, it is abnormal excessive brain activity; and for psychogenic TLOC it is the psychological process of conversion (Figure 1).

Syncope is defined as TLOC due to cerebral hypoperfusion, characterized by a rapid onset, short duration, and spontaneous complete recovery. Global cerebral hypoperfusion occurs when systemic blood pressure (BP) falls below a certain limit which varies with age in the paediatric population. Systemic BP is the product of cardiac output and total peripheral resistance; a fall in either can cause syncope. However, in syncope, both mechanisms often act together to a varying degree (Figure 2).

Syncope is very common in the paediatric population and up to 15% of children can experience syncope by the end of adolescence.

The vast majority of paediatric syncope does not have is a cardiac cause.

The risk of sudden death in fainting children is extremely small.

However, in a very small number of patients it can be the first presentation with something more serious. It is therefore very important to assess red flag symptoms for high risk patients that need to be referred to Paediatric Cardiology services.

NB: A fraction of patients diagnosed with epilepsy actually have an underlying arrhythmogenic substrate; there is increasing evidence that some of the channelopathies can have a mixed phenotype with neurological and cardiac symptoms, therefore paediatric neurologist and paediatrician need to be aware of the relevant presentations and conditions.

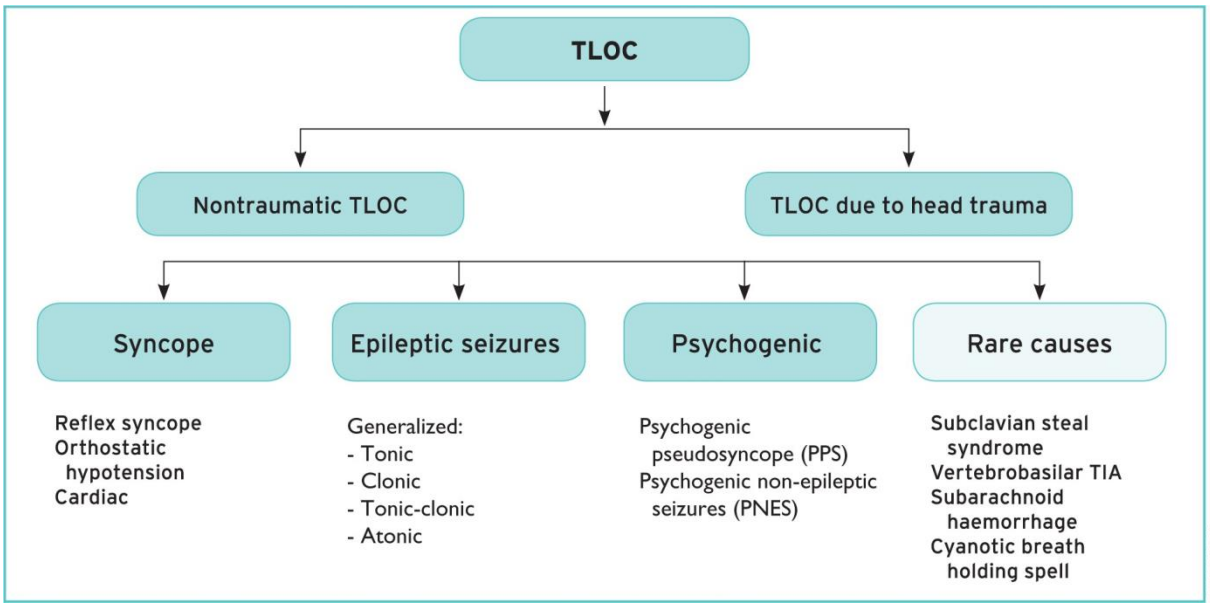


Figure 1: Syncope in the context of transient loss of consciousness.
Adopted from ESC 2018 guidelines [(1)]

2.1 Main causes of syncope

Reflex (neurally mediated) syncope

- 'Pallid syncope'; usually in toddlers

- Vasovagal: usually has prodromal symptoms
    - orthostatic vasovagal syncope: standing, less common sitting
    - emotional: fear, pain (somatic or visceral), instrumentation, blood phobia (however LQT syndrome can be triggered by fright, anger or loud noises)

- Situational:
    - micturition
    - gastrointestinal stimulation (swallow, defaecation)
    - cough, sneeze
    - post-exercise
    - others (e.g. laughing, brass instrument playing)

- Carotid sinus syndrome

- Non-classical forms (without prodromes and/or without apparent triggers and/or atypical presentation)

Orthostatic hypotension (OH)

Note that hypotension may be exacerbated by venous pooling during exercise (exercise-induced), after meals (postprandial hypotension), and after prolonged bed rest (de-conditioning).

- Teenage 'dysautonomia'; immaturity of the autonomic control around puberty
- Drug-induced OH (most common cause of OH):
    - e.g. vasodilators, diuretics, phenothiazine, antidepressants
- Volume depletion:
    - haemorrhage, diarrhoea, vomiting, etc
- Primary autonomic failure (neurogenic OH):
    - pure autonomic failure, multiple system atrophy
- Secondary autonomic failure (neurogenic OH):
    -Diabetes, spinal cord injuries, auto-immune autonomic neuropathy, paraneoplastic autonomic neuropathy, renal failure

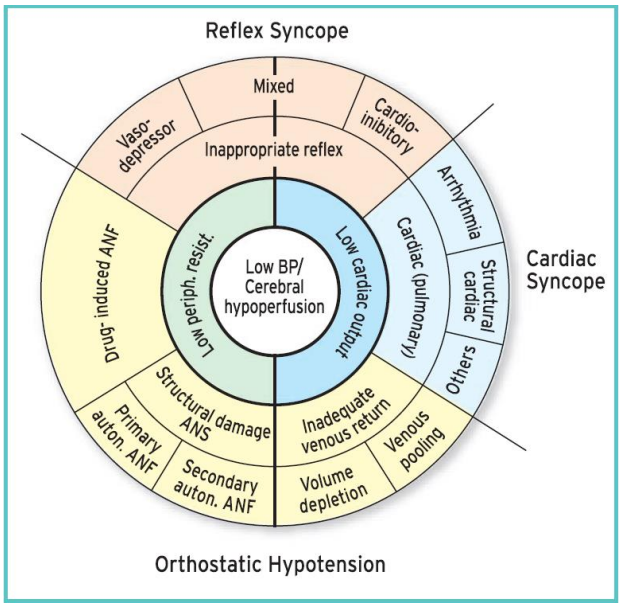


Figure 2. Pathophysiological basis of syncope classification.
ANS = autonomic nervous system, auton. = autonomic, BP =blood pressure, OH= orthostatic hypotension, periph. =peripheral, resist. = resistance. Adopted from ESC 2018 guidelines [(1)].

2.2 Cardiac syncope; a rare but anxiety provoking cause of syncope in children

2.2.1 Causes

- Arrhythmia as primary cause:
    - Bradycardia (quite rare in the paediatric population)
      - sinus node dysfunction
      - atrioventricular conduction system disease
    - Tachycardia:
      - supraventricular
      - ventricular (often channelopathy related)
- Structural cardiac causes are rare, but if syncope is present in children with known congenital heart disease, it must be investigated carefully:
    - congenital heart disease (especially left ventricular outflow obstruction), hypertrophic cardiomyopathy,
    - congenital or acquired coronary artery anomalies (previous Kawasaki disease)
    - prosthetic valve dysfunction.
- Rare even in children: choking games, pericardial disease/tamponade (usually cardiac post-op or myo-/pericarditis), acute myocardial ischaemia (usually in the context of viral myocarditis), acute aortic dissection (Marfanoid/dysmorphic features), pulmonary hypertension (usually ex-preterm with severe chronic lung disease), pulmonary embolus.

2.2.2 Clinical assessment

i. History

- **Detailed history is the cornerstone in syncope differential diagnosis (3).**
- First aim is to establish whether there was indeed a TLOC and can be classified as syncopal episode (short duration <30 sec, abnormal motor control, loss of responsiveness and amnesia for the period of LOC).
- The age of the child is paramount; syncope in a teenager or a toddler have significantly different implications to syncope in a 7 year old.
- Assessment of the situational settings and what has occurred before, during and after the TLOC is paramount. Seizures may be a sequelae of syncope.
- A video recording is often invaluable to understand the underlying cause.
- Medications, including vasoactive substances such as diuretics, and other drugs (including of abuse) should be assessed.
- Family history with detailed family tree is also very important and should include unexplained accidents and deaths in extended family members as well as use of any implantable device.
- **The questions asked should actively exclude the presence of red flags (Figure 3 and Syncope Referral Guideline on page 2):**

Figure 3: Risk stratification for underlying cardiac disease

Risk stratification for underlying cardiac cause				
	Presentation	**Past medical history**	**Examination**	**ECG**
Low risk	Associated with prodrome typical of reflex syncope[1] After sudden unexpected unpleasant sight, sound, smell or pain After prolonged standing or crowded, hot places During meals or postprandial Triggered by cough, defaecation or micturition With head rotation or pressure on carotid sinus Changing position Fatigue following the event	Long history of recurrent syncope with low risk features with the same characteristics as the current episode Absence of structural heart disease	Normal examination	
High risk	Syncope during exertion (especially during swimming) or when supine Sudden onset palpitations immediately followed by syncope Short duration (<30sec) Quick recovery No warning symptoms or short prodrome (<10sec) Syncope in the sitting position Sensorineural deafness (LQTS)	Structural heart disease Heart failure Ventricular dysfunction Previous treatment for Kawasaki syndrome Previous history of "near syncope" (has the same prognosis as syncope) Family history of SCD at young age	Pathological heart murmur Organomegaly Failure to thrive Abnormal observations (especially fixed tachycardia) in the absence of underlying cause	High degree AV block [2] Inappropriate for age tachycardia with fixed heart rate SVT or VT Type 1 Brugada pattern (Figure 1) ECG changes consistent with ischemia Preexcitation Long QTc or short QTc (<360msec) Figure @@ Negative T waves in right precordial leads in teenagers (beyond V2) Partial RBBB in teenagers Epsilon wave (Figure @@ ARVC) Left or right ventricular hypertrophy Patients with pacing devices Frequent ventricular ectopics [3] Persistent sinus bradycardia or sinus pauses >3 s in awake state and in absence of physical training

[1] light headiness, pallor, feeling of warmth, visual disturbances, sweating, nausea, vomiting

[2] complete heart block, Mobitz II second degree AV block, markedly prolonged PR with symptoms

[3] >1 per ECG strip or during auscultation; possible ARVC, catecholaminergic polymorphic VT

Figure 3. High-risk features (that suggest a serious condition) and low-risk features (that suggest a benign condition) in patients with syncope at initial evaluation.
Modified from ESC 2018 guidelines (1).

i. Physical examination:

- Cardiac examination, looking for murmurs and evidence of heart failure (signs of respiratory distress, failure to thrive, organomegaly).
- Brief age-appropriate neurological examination.
- Look for evidence of injury sustained as a result of the syncopal episode.

ii. Observations:

- Vital signs including BP.
- Lying and standing BP and HR – abnormal if HR increases by 20 bpm when standing from sitting, or systolic BP decreases by 10mmHg (suggesting a circulatory cause / orthostatic hypotension).
- Check the femoral pulses; Upper vs. lower limb blood pressure – a gradient of >20mmHg is suggestive of coarctation
- Murmurs?

iii. ECG:

- **Most patients with syncope need an ECG (See Paediatric ECG guideline).** However, if the history is very clear (age, situation, witnessed etc) that there is a non-cardiac cause eg vasovagal or orthostatic syncope then an ECG is unnecessary, and can be anxiety provoking if 'normal variants' such as sinus arrythmia are present.
- When there is Syncope without a clear history or in an unusual age-group (2 – 10) or and unusual situation an ECG should be performed
- A normal ECG is reassuring but does not always exclude a cardiac cause (Figure 4).

iv. Ambulatory ECG

- The diagnostic yield is usually low and depends on the event frequency. Event recorder is recommended for syncope and Holter monitoring for palpitations. Arrhythmic syncope is confirmed when a correlation between syncope and an arrhythmia is detected.

v. Laboratory tests

- Consider blood tests e.g. FBC, U&E, TFT when a specific cause is suspected or before referring to cardiology to exclude non cardiac causes.
- Consider blood sugar level if patient seen shortly after event +/- symptoms of hypoglycaemia.
- Pregnancy test for all female teenagers.

- Consider urine toxicology in patients with prolonged decreased consciousness or confusion.

Exercise testing is indicated especially for patients who experience syncope during or shortly after exertion.

Patients who have a cardiac device and syncope should undergo prompt device interrogation.

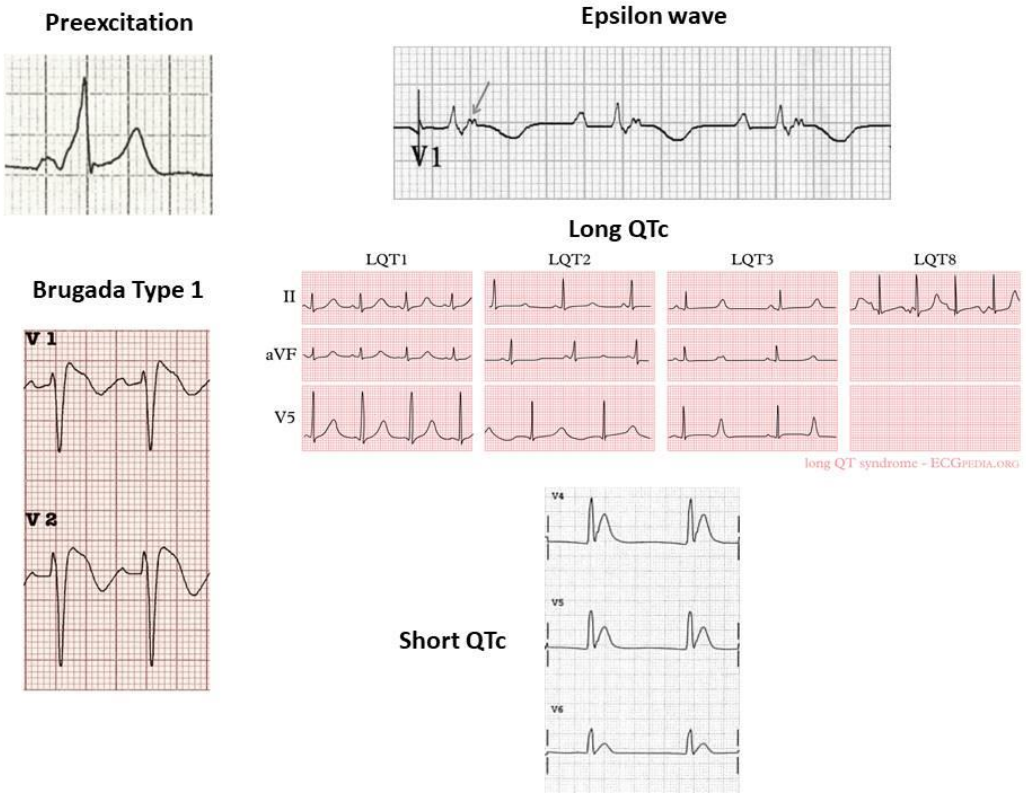


Figure 4. ECG findings of concern requiring Cardiology Referral
(Adopted from https://litfl.com/ https://en.ecgpedia.org)

3. Risk management

There are three main questions to be answered when trying to risk stratify patients with TLOC.

1. Is there a serious underlying cause that can be identified?
2. What is the risk of a serious outcome?
3. Should the patient be admitted to hospital?

4. Advice

The majority of patients will have a benign underlying cause of syncope and can be discharged with safety net advice.

- The absence of red flags with a normal cardiac examination (and normal ECG if undertaken) indicates a low risk for cardiac cause of syncope. If any red flag features mentioned occur seek medical attention.
- Ensure adequate oral fluid and salt intake, avoid caffeine, and make some behavioural changes:
    - don't stand up too quickly from seated position
    - keep knees slightly bent when standing for long periods
    - move your toes frequently if you are standing / sitting for some time
- Drink 2 litres a day if less than 10 years and 2.5 litres a day if above 10 years of age; and salt supplements up to 5g (1 teaspoon) a day
- Seek medical advice if chest pain, palpitations or significant SOB develop, if symptoms recur with no obvious trigger, last longer than a few seconds or if confused for longer than 20 minutes after the episode.
- Parents can be pointed towards relevant websites for further information in benign causes of syncope.
    - Take Fainting to Heart - STARS - UK (heartrhythmalliance.org)

5. Education and Training

It takes a minimum of 5 years post MRCP/MRCPCH (ie ST4+) training to become a Paediatric Cardiologist. Paediatric (Congenital and Paediatric) Cardiology is a full 'physicianly' speciality. Paediatric Cardiology at UHL is part of the East Midlands Congenital Heart Centre (EMCHC) now split with the paediatric centre in the Kensington building at LRI and the Adult Congenital Heart Disease (ACHD) centre above the South Entrance at Glenfield. 'Paediatric Cardiologists' can train in a wide range of subspeciality areas from fetal cardiology, across childhood and into the care of adults born with CHD, and includes for example complex structural interventions, electrophysiology, and the use of state of the art imaging modalities. Adult cardiologists can also subspecialise in ACHD. EMCHC is the recognised training centre for the East Midlands and currently trains 4 NTNs in Paediatric Cardiology, I – 2 NTNs in ACHD, as well as 1-2 Paediatricians with expertise in Cardiology (SPIN module from Paediatrics at ST6-8) and a number of fellows. The specialists work hand in hand with Congenital heart surgery, intensive care and specialist anaesthesia, as well as relying on many other specialists for their many patients with complex medical needs.

6. Monitoring Compliance

None

7. Supporting References

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   2017 ACC/AHA/HRS guideline for the evaluation and management of patients with syncope: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. *Circulation*. 2017;136:e60–e122. DOI:10.1161/CIR.0000000000000499
3. MacCormick JM, McAlister H, Crawford J, et al. Misdiagnosis of long QT syndrome as epilepsy at first presentation. Ann Emerg Med 2009;54:26–32.
4. Harris M, Bu'Lock F. Fifteen-minute consultation on limiting investigations in the fainting child. Archives of Disease in Childhood - Education and Practice 2016;101:26-30.

8. Key Words

Syncope, fainting, Chest pain, palpitations, exercise,

The Trust recognises the diversity of the local community it serves. Our aim therefore is to provide a safe environment free from discrimination and treat all individuals fairly with dignity and appropriately according to their needs.
As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.

CONTACT AND REVIEW DETAILS			
Guideline Lead (Name and Title) Prof Frances Bu'Lock - Cardiologist			**Executive Lead** Chief Medical Officer
Details of Changes made during review:			
Date	**Issue Number**	**Reviewed By**	**Description Of Changes (If Any)**
March 2023	**3**	**F Bu'Lock** **S Shebani** **Paediatric Cardiology Consultants group** **UHL Children's services quality & safety board**	**Complete overhaul of original document**